

# PEDIATRIC NEUROLOGY BRIEFS

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J. GORDON MILLICHAP, M.D., F.R.C.P., EDITOR

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### PAROXYSMAL DISORDERS

#### PROSPECTIVE STUDY OF BREATH-HOLDING SPELLS

The natural history of severe breath-holding spells (BHS) was studied prospectively in 95 children (48 boys, 47 girls) referred for neurologic consultation at Children's Medical Center, Hartford, CT. A structured interview was conducted initially and at 1-year intervals. A positive family history of BHS in a near relative was present in 34%, of equal frequency on paternal and maternal sides. The cyanotic type of BHS was reported in 52%, the pallid type in 28%, and both types occurred in 20%. Electrocardiogram recordings performed on children with pallid spells were all normal. Median age of onset of BHS was 6 to 12 months, and 15% presented younger than 6 months. Four patients (5%) had a first attack as a neonate, at less than 24 hours to 29 days of age, and only 3 had a late onset, between 25 and 30 months. The median frequency of spells was daily to weekly, the average being weekly, and 25% had >1 per day at peak frequency. The age at peak frequency was 13 to 24 months with a mean of 18 months. The median age at termination of spells in 67 patients was 37 to 42 months; 15 children had the last attack after age 4 years, the oldest at 7 years of age. Hypoxic convulsions occurred in association with the BHS in 15%. Phenobarbital and phenytoin administered in 3 failed to control the seizure. Syncopal episodes precipitated by trauma or stress occurred in 12 (18%) of 67 patients who were followed after BHS had terminated. (DiMario FJ Jr. Prospective study of children with cyanotic and pallid breath-holding spells. *Pediatrics* February 2001;107:265-269). (Reprints: Francis J DiMario Jr MD, Department of Pediatrics, Connecticut Children's Medical Center, 282 Washington St, Hartford, CT 06101).

**COMMENT.** Breath-holding spells (BHS) are an involuntary, reflexic, nonepileptic paroxysmal disorder of infancy and early childhood. They are characterized by the following sequence of symptoms: 1) a precipitating factor such as slight injury or emotional upset; 2) crying of short duration; 3) a respiratory gasp and breath holding in expiration; 4) cyanosis and/or pallor; 5) opisthotonic rigidity and loss of consciousness; and 6) flaccidity or convulsive movements. Three types of BHS are described, cyanotic, pallid, and mixed

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(Lombroso CT, Lerman P. Pediatrics 1967;39:563-581). However, the absence of cyanosis in BHS is disputed by some authorities (Livingston S. Comprehensive Management of Epilepsy in Infancy, Childhood and Adolescence. Springfield, IL, Charles C Thomas, 1972). So-called *pallid BHS* are precipitated by trauma rather than anger, and are associated with cardiac asystole and EEG slowing. The pathophysiology of the pallid BHS is vagal cardiac inhibition and cerebral anoxia, and treatment with atropine may be beneficial. Under carefully controlled EEG and cardiac monitoring, the diagnosis has been confirmed by ocular compression (Brenningstall GN. Pediatr Neurol 1996;14:91-97). A fall in arterial oxygen saturation has been demonstrated during an attack (Gauk EW et al. N Engl J Med 1963;268:1436-1441). An iron deficiency anemia found in 23% of cases of BHS is suggested as a possible contributing causative factor (Holowach J, Thurston DL. N Engl J Med 1963;268:21-23; Ped Neur Briefs May 1997;11:33). Autonomic dysregulation is a common mediating mechanism for both pallid and cyanotic BHS, resulting in loss of consciousness (DiMario FJ Jr, Burleson JA. Pediatr Neurol 1993;9:268-274).

As demonstrated in the above study and in many previous clinical reports, although a BHS is a frightening disorder for parents, the prognosis is invariably benign, attacks usually ending by age 5 years. The differential diagnosis includes a cardiac pathology with prolonged QT interval or an epilepsy. An EKG is recommended in children with pallid BHS, and an EEG in those with prolonged convulsive movements. In BHS the EEG is normal and treatment with conventional antiepileptic drugs is ineffective. Treatment requires parental counseling: 1) frequent reassurance about the benign nature of the spells, 2) an explanation of the strong genetic factor, with an autosomal dominant inheritance and equal male to female ratio, and 3) the risks of a tendency to syncopal attacks and behavior disturbance in later childhood. A trial of iron therapy in patients with a lowered hemoglobin level may be beneficial. Of 33 children treated with ferrous sulfate orally (5 mg/kg/day for 16 weeks) 88% had a complete or partial control of BHS, whereas in 34 receiving placebo only 6% resolved (Daoud AS et al. J Pediatr 1997;130:547-550; Ped Neur Briefs 1997;11:33).

## SEIZURE DISORDERS

### **RISK OF EPILEPSY AFTER COMPLEX FEBRILE SEIZURES**

The risk of epilepsy developing after complex febrile seizures (CFS) was studied in 477 children admitted between 1991 and 1998 with febrile convulsions at Tel Aviv Medical Center, Israel. Among 57 (12%) diagnosed with CFS and an otherwise normal neurologic exam, 48 were available for follow-up, and 13 (27%) developed epilepsy. Thirty percent had a family history of febrile seizures, but none had a positive family history of epilepsy. The follow-up period was 8-78 months (mean 43 +/- 24 months). Among patients developing epilepsy, the mean age of CFS onset was 11 months compared to 17 months in those without epilepsy. CFS of the partial seizure type had a higher risk of epilepsy (45%) than those with multiple febrile seizures (21%). None of 3 patients with the prolonged type of CFS developed epilepsy at follow-up. Neuroimaging studies performed in 15 patients were all normal, and 10 had partial CFS. Two of the 13 patients with epilepsy treated with anticonvulsants were refractory. (Sapir D, Leitner Y, Harel S, Kramer U. Unprovoked seizures after complex febrile convulsions. Brain Dev Dec 2000;22:484-486). (Respond: Dr Uri Kramer, Child Development Center, Beit Habriut Straus, 14 Balfour Street, Tel Aviv 65211, Israel).